Case reports

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Pulmonary atresia with intact ventricular septum *Open heart surgical correction at 32 hours*

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A case of pulmonary atresia with intact ventricular septum is reported in which total surgical correction was carried out successfully at 32 hours of age. Cardiac catheterisation at 17 months has revealed virtually normal haemodynamic and angiographic findings. Surgical correction of this condition in the neonatal period is discussed.

When the pulmonary valve is atretic and the ventricular septum is intact, systemic venous return enters the left atrium via a patent foramen ovale or atrial septal defect. The lungs are perfused by aortopulmonary collateral vessels or a persistent ductus arteriosus, and hypoxia is frequently severe. Patients present in the early neonatal period with severe right heart failure and cyanosis. There may be a systolic murmur of tricuspid regurgitation or continuous murmur of a persistent ductus arteriosus and the second heart sound is single.

The chest radiograph shows oligoemic lung fields and cardiac enlargement with a prominent right atrium. The electrocardiogram shows an inferiorly directed mean frontal QRS axis, tall P waves in lead II, and either a low R/S ratio in lead V1 or right ventricular hypertrophy. The echocardiogram is not very helpful but the pulmonary valve is not seen. Cardiac catheterisation and angiocardiography are diagnostic.

In most cases the main pulmonary artery is patent and therefore total correction or palliative surgery can usually be attempted.

Case report

The patient was a full-term normal infant weighing 4.05 kg and was centrally cyanosed at birth. At 24 hours there was severe cyanosis with right heart failure, a pansystolic murmur at the left sternal edge, and single second heart sound. The electrocardiogram showed tall P waves in lead II, and R/S ratio of 0.8 in lead V1, and a mean frontal QRS axis of +90°. The chest radiograph (Fig. 1a) showed

oligoemic lung fields and cardiac enlargement with a prominent right atrium. At right heart catheterisation with selective right (Fig. 2a) and left ventriculography, the appearances were those of pulmonary atresia with intact ventricular septum, patent foramen ovale, and persistent ductus arteriosus. The right ventricle was hypoplastic with pronounced infundibular narrowing and there were multiple sinusoids communicating with the coronary veins. The main pulmonary trunk, filling via the persistent ductus arteriosus, was of good size and extended proximally to an atretic pulmonary valve. Balloon atrial septostomy was performed.

At operation (L.D.A.) at 32 hours of age, the patient was surface cooled to 25°C with 'Cryogel' packs and then to 18.5°C using extracorporeal circulation. Deep hypothermia with circulatory arrest was maintained for 26 minutes. The persistent ductus arteriosus was ligated. The main pulmonary artery was opened revealing an imperforate pulmonary valve with three normal-sized cusps. The fused commissures were incised. Infundibular muscle was resected through a transverse right ventriculotomy and the atrial septal defect was closed through a right atriotomy. The patient developed severe right heart failure in the immediate postoperative period, but this gradually responded to diuretics, though treatment with frusemide was required until the age of 5 months.

At 17 months she was thriving and was developing normally. There was then a short systolic ejection murmur at the left sternal edge, a normally split second heart sound, and an early diastolic murmur in the second left intercostal space. The

Fig. 1 (a) Preoperative chest radiograph showing cardiac enlargement, right atrial enlargement, and oligemic lung fields. (b) Postoperative chest radiograph at 17 months showing slight cardiac enlargement.

Fig. 2 (a) Preoperative right ventriculogram showing severe tricuspid regurgitation, multiple intramyocardial sinusoids, and a blind outflow tract. (b) Postoperative right ventriculogram showing normal-sized right ventricle and pulmonary arteries, with trivial tricuspid regurgitation.

chest x-ray (Fig. 1b) showed slight cardiac enlargement with a prominent right atrium and normal vascularity of the lung fields. The electrocardiogram showed a mean frontal QRS axis of $+120^\circ$, but was otherwise normal. At right heart catheterisation (Table), selective right ventricular angiography (Fig. 2b) showed a large trabeculated right ventricle with a wide outflow tract leading to a normal sized pulmonary artery, and a trace of regurgitation through a normally situated tricuspid valve. There was no evidence of a residual atrial septal defect on angiography or oximetry and the catheter would not pass from the right to the left atrium.

Discussion

Pulmonary atresia with intact ventricular septum has been classified into 2 groups, according to the size of the right ventricular cavity (Greenwold *et al.*, 1956; Davignon *et al.*, 1961). It has been thought that type II (large right ventricle) was more amenable to correction by pulmonary valvotomy and carried a better prognosis than type I (hypoplastic right ventricle). It seems that there is a spectrum of severity in those patients with hypoplasia of the right ventricle (Gersony *et al.*, 1967; Bowman *et al.*, 1971; Miller *et al.*, 1973) but intramyocardial

Table Postoperative pressures (at 17 months)

Right atrium (mmHg)	Right ventricle (body) (mmHg)	Right ventricle (outflow) (mmHg)	Pulmonary artery (mmHg)	Pulmonary artery wedge (mmHg)	Femoral artery (mmHg)
a=9, x=6 v=9, y=6 (mean 7)	26/5 (end diastolic 10)	26/5 (end diastolic 10)	19/8 (mean 11)	(mean 10)	84/50 (mean 66)

sinusoids can almost always be demonstrated (Lauer *et al.*, 1964). Our patient had a hypoplastic right ventricle with intramyocardial sinusoids (type I). It is significant that after operation the right ventricle grew to a normal size and the tricuspid regurgitation became trivial. We are not aware of any other reports of total correction at such an early age.

The place of pulmonary valvotomy for neonates with pulmonary atresia, intact ventricular septum, and varying degrees of hypoplasia of the right ventricle is debatable (Greenwold *et al.*, 1956; Vlad, 1973). There have been reports of survival after pulmonary valvotomy in patients with hypoplastic as well as in those with large right ventricles (Gersony *et al.*, 1967; Celemajer *et al.*, 1968; Cole *et al.*, 1968; Dhanavaravibul *et al.*, 1970; Bowman *et al.*, 1971; Murphy *et al.*, 1971; Shams *et al.*, 1971), but it has been suggested (Miller *et al.*, 1973) that some of these reports may have included patients with critical pulmonary stenosis, the prognosis of which is much better. Shams and co-workers (1971) reported 7 survivors of 8 patients in whom aortopulmonary shunts were performed and atrial septal defects created. Rook and Gootman (1971) reported 3 cases of type II pulmonary atresia who survived pulmonary valvotomy, though 2 patients required an additional shunt operation.

The conventional approach to operation for pulmonary atresia with intact ventricular septum is probably that advocated by Vlad (1973). He recommends balloon atrial septostomy (Miller *et al.*, 1969) and aortopulmonary shunt for all patients with a hypoplastic right ventricle, but pulmonary valvotomy when the ventricle is large. With this approach he reports a 44 per cent survival. It is clear, however, that some patients with type I pulmonary atresia do well after valvotomy and that the right ventricle may grow normally, and that pulmonary valvotomy will not always be successful in patients with a large right ventricle (type II). Our patient underwent open pulmonary valvotomy with outflow tract resection, and this more extensive procedure may have resulted in greater haemodynamic improvement, allowing the right ventricle to grow after operation.

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